A Multidisciplinary Approach to Paediatric Hearing Loss: Programme at the Centre for Hearing Intervention and Language Development, National University Hospital, Singapore

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Abstract

The objective is to describe the multidisciplinary management programme at the National University Hospital (NUH) in Singapore for children with hearing impairment (HI). Over 99.95% of babies born at NUH have hearing tested with both otoacoustic emission and automated auditory brainstem response tests by 6 weeks of age. The referral rate to Otolaryngology is 0.5%. Acquired causes of congenital HI are decreasing. Thirty percent of patients at NUH with idiopathic congenital sensorineural HI have DFNB1/ GJB6 Connexin 26 HI. CT scan or MRI imaging has a higher diagnostic yield when there is unilateral, fluctuating or non-Connexin 26 related HI. Routine electrocardiogram and Opthalmology evaluations will exclude associations of fatal cardiac rhythm anomaly and retinopathy. Other investigations are directed by history and clinical examination. There is now a very wide range of increasingly sophisticated medication, neuro-otologic external, middle and inner ear surgery, hearing aids, middle ear implants and cochlear implants available to improve hearing. A multidisciplinary team from neonatology, paediatrics, otolaryngology, audiology, auditory verbal and speech therapy, ophthalmology, radiology, and psychology working closely with the child, family and schools is needed to develop a cost-effective and comprehensive management programme for paediatric HI.

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Introduction

Congenital hearing impairment (HI) occurs in approximately 4 per 1000 newborns worldwide. If intervention is delayed, the child with HI will have delayed speech and language acquisition, impaired cognition, and difficulty integrating into mainstream society socially and at work. I was asked to present on the multidisciplinary management programme at the National University Hospital (NUH) in Singapore currently available for children with HI.

Detection of Paediatric Hearing Loss

Universal Newborn Hearing Screening (UNHS) is performed on approximately 99.95% of newborns delivered at NUH within 6 weeks of life.[1] Both Otoacoustic Emission (OAE) and Automated Auditory Brainstem Response (ABR) hearing tests are administered by colleagues from the Neonatology Department. Those failing UNHS screening are referred to Otolaryngology. Paediatric otolaryngologists first exclude ear conditions that need medical or surgical interventions. Paediatric audiologists then perform tympanogram, OAE, ABR and steady state evoked potential (SSEP) hearing tests. Hearing aids can be fitted as early as 1.5 months if appropriate. The referral rate to ENT from UNHS was 0.5% (33/6836) for newborns between 2003 and 2005. Among these 33 newborns, 4 had false-positive HI, 3 defaulted follow-up and 26 had HI.

Age-appropriate hearing tests include behavioural observation audiometry for children less than 3 years old, play audiometry for children 3 to 5 years old, and pure tone audiometry from 5 years old. ABR is used in neonates and for children unable to cooperate with subjective testing when awake.

Missing a Diagnosis of Hearing Impairment

HI is easily missed. A child with mild-moderate HI can still hear door bells and telephones, and can use visual and other sensory cues to aid in understanding what caregivers are saying. For normal development of speech and language however, near perfect hearing is needed. Many consonants of speech are heard only at 20 to 30 dB levels. Even mild HI may therefore result in speech, language, cognition and behavioural disorders. Only a formal hearing test can detect

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mild HI accurately. HI is easily missed with informal testing in a non-sound proof room. Hearing tests need to be repeated if there is any suspicion of HI. A genetic or congenital HI may pass UNHS but manifest later in childhood, or progressively deteriorate or fluctuate.

In Central Auditory Processing Disorder (CAPD), hearing tests are normal, but decoding of auditory information is impaired. Auditory sound information may not be optimally heard by the child, especially in challenging situations of background noise, poor room acoustics, rapid speech or long sentences. Special speech lists testing are needed for diagnosis. The centre started comprehensive CAPD evaluation and tailored CAPD management in Singapore between 2004 and 2005. We have found that definitions of norms used in the West may not be appropriate for Singapore, so we are developing Singapore-specific CAPD normative data for Singaporean children.[2] After the medical evaluation by paediatric otolaryngologists, paediatric audiologists sub-specialising in CAPD administer the CAPD tests, and a speech language therapist sub-specialising in CAPD works with the family and child on a tailored programme of rehabilitation strategies.

Excluding an Infectious Cause of Deafness

In NUH, every newborn is screened for hypothyroidism at birth. Infectious causes of HI due to TORCHES (Toxoplasmosis, Rubella, Cytomegalovirus, Herpes, Syphilis) infections are decreasing. Toxoplasmosis and Herpes Simplex infections are also uncommon with improved hygiene and standards of living. The cost-effectiveness of universal neonatal screening for Cytomegalovirus (CMV) is currently controversial. However, a diagnosis of congenital CMV can only be made if tested for within the first month of life, and CMV accounts for 10% to 35% of congenital HI in the West. HI in congenital CMV affects 40% to 60% of symptomatic infants and 7% to15% of asymptomatic infants. Both HI and neurological deterioration can progress undetected in asymptomatic CMV. HI due to CMV can develop months or even years after birth.[3] Early ganciclovir therapy may reverse neuro-developmental delay and HI.[4]

Excluding a Genetic Cause of Deafness

Genetic testing for congenital HI is cost-effective, and is now performed worldwide.[5] Seventy-five percent of genetic HI is non-syndromic, and 75% of non-syndromic HI is autosomal recessive in inheritance. Babies with genetic HI therefore look normal, are well except for HI, and are born to hearing parents. It is thus easy to miss a diagnosis of genetic HI unless a blood test of DNA is done. The most common genetic mutation responsible for non-syndromic, autosomal recessive HI is a DFNB1/ GJB2 mutation that results in an abnormal Connexin 26 gap junction protein required for potassium recycling in the cochlear. Connexin 26 HI alone accounts for 20% to 60% of idiopathic congenital sensorineural HI in the West.[6,7] In a recently concluded ASTAR-BMRC funded study evaluating idiopathic HI, Connexin 26 HI was responsible for 3 out of 10 cases of idiopathic congenital sensorineural HI seen at NUH.[8] It is thus now the first test in our evaluation of the etiology of idiopathic congenital bilateral sensorineural HI. Singapore's most prevalent connexion 26 mutation is the missense mutation V37I, unlike the 35delG and 235delC nonsense mutations common in the West and China respectively. Connexin 26 HI ranges from mild to profound in severity and can progress with time. No A1555G mitochondrial mutations which increase the risk of aminoglycoside-induced HI, nor Connexin 30 (DFNB2/ GJB6) mutations were identified in our Singapore population. Genetic testing for connexin 26, connexin 30 and A1555G mitochondria congenital HI was made available in 2004 as a diagnostic service in NUH at the Molecular Diagnostic Laboratory.

Excluding Other Causes of Deafness

Urinary blood and protein analysis may alert us to the renal dysfunction associated with HI in Alport's syndrome. Electrocardiogram is a simple and inexpensive investigation to exclude the prolonged QT interval associated with Jervell Lange Nielsen (JLN) syndrome, especially important if there is a family history of sudden death or syncope. Eye examination is especially important in those with severe-profound HI as 50% of these children may have concomitant visual problems, example like in Usher's Syndrome. Early retina treatment can prevent or delay the blindness. CMV and Toxoplasmosis retinitis can also be identified. Co-management with geneticists and multidisciplinary paediatric specialists are important in children with syndromes affecting multiple systems. Children with cleft palates and craniofacial anomalies are co-managed with plastic, maxillofacial and orthodontic surgeons. Syndromic HI may have mixed conductive and sensorineural HI. If conservative management fails, conductive HI may benefit from insertion of ventilation middle ear tubes, repair of obvious or occult cleft palates, replacement of middle ear ossicles, and mastoid surgery to remove cholesteatoma tumour and chronic mastoid infection.

Radiological imaging gives higher diagnostic yield than genetic tests in children with unilateral or fluctuating HI.[9] Enlarged vestibular aqueduct (EVA), cochlear and semicircular canal anomaly in the temporal bone can be detected on computer tomography (CT) scan. In a recently completed study on patients with EVA seen at NUH, our patients had larger overall mean diameter of EVA, and higher association with Mondini's cochlear compared to those in reported literature.[10] Most had late diagnosis and

severe-profound bilateral hearing loss that needed hearing aids or cochlear implants. If EVA is diagnosed, genetic testing for Pendrin is useful to exclude Pendred disease in which there is EVA, possible cochlear malformation, and thyroid dysfunction. Children with EVA should avoid sudden increase in intracranial pressure that may cause deterioration in hearing (examples are head trauma, diving and contact sports). Early steroid therapy may reverse sudden deterioration in hearing in EVA. Magnetic resonance imaging (MRI) can detect an absent or hypoplastic cochlear nerve, cochlear fibrous ossification and brain pathology better than the CT scan.

Other investigations ordered are guided by history, examination, and careful follow-up of the child. For example, post-lingual progressive HI may be associated with autoimmune problems, and HI will be associated with branchial fistulas and renal dysfunction in Branchio-Oto-Renal Syndrome.

Hearing and Speech Rehabilitation

Children are fitted with appropriate hearing aids (HA) as early as 6 weeks old by our paediatric audiologists. A wide range of sophisticated digital HAs with greatly improved sound quality and cosmesis is now available. Behind-the-ear HAs are usually used for growing children as they require frequent change of ear moulds. When HAs fail to give optimal hearing outcomes, a similarly wide range of hearing implants and middle ear reconstruction surgery is now available. Malformed ears can be managed with silicon prosthetic ears, and reconstruction of the auricles. HI due to ear canal atresia and congenital anomaly of the middle ear bones can be managed with bone-anchored hearing aid, canalplasty, ossiculoplasty and middle ear implant surgery. The centre performed Asia's first Vibrant Soundbridge middle ear implant (MEI) on the incus for adult presbyacusis in 2006, and expanded MEI use to round and oval window applications for mixed or conductive HI in 2007. In 2008, the centre performed the first totally implantable Carina MEI in Singapore for moderate congenital sensorineural HI in an adult who was not satisfied with HAs. Severe to profound bilateral HI may require cochlear implants. The centre pioneered simultaneous bilateral cochlear implantation in 2006 in Singapore. Bilateral hearing can translate to better speech and language acquisition outcomes, ease of listening in background-noise situations, and improved localisation of sound. The selection of an appropriate candidate is made only after thorough medical, audiology and auditory verbal rehabilitation assessments, with pros and cons fully discussed with the family.

Auditory Verbal Therapy (AVT) rehabilitation at our centre is done by Speech Language Therapists who further undergo AVT training. The aim is to integrate the child with HI into mainstream society, without the need for lip reading and signing to communicate if possible. Rehabilitation is tailored to the child and family, with some patients performing better with some cued speech, especially if they have additional problems like autism and cognitive delay. Children may also have speech language impairment and other language problems coexisting with HI that needs to be identified and co-managed with other therapists. A holistic and coordinated rehabilitation programme tailored to the child is ideal. It often involves the otolaryngologist, speech and language therapist, physiotherapist, occupational therapist, specific paediatric medical specialist, psychologist, medical social worker and teachers from the child's school (main-stream, Canossian School or School for the Deaf).

Conclusion

A multidisciplinary team from neonatology, paediatrics, paediatric otolaryngology, paediatric audiology, speech language and auditory verbal therapy, child development, genetics, ophthalmology, radiology, medical social work, and psychology working closely together with the child, family and schools is needed to develop a cost-effective and comprehensive management programme for paediatric HI.

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